

(EP05.104).Vascular limb marvel in utero: Klippel-Trenaunay-Weber syndrome unveiled

Dr. Ladbans Kaur, Dr. M Preet, Dr. A Bhalla

Case Report : Klippel-Trenaunay Syndrome (KTS) is a rare congenital condition defined by a triad of capillary malformations (port-wine stains), venous malformations, and overgrowth of soft tissue and bone. A 27-year-old primigravida presented at 18 weeks of gestation. Ultrasound examination revealed a diffuse heterogeneous lesion with tortuous anechoic channels involving the subcutaneous tissues of the right lower limb, including the thigh, leg, and dorsum of the foot (Fig 1a, b, c). The affected limb was significantly oedematous and longer due to bone hypertrophy compared to the normal left lower limb (Fig 2). Power Doppler imaging demonstrated spontaneous flow within the dilated anechoic channels and spectral doppler showing continuous flow indicative of venous lacunae due to haemangiomas (Fig 3). Despite the anomalies, the lengths of the femur, tibia, and fibula in the right lower limb were appropriate for the gestational age. Additionally, fetal echocardiography was normal. No other abnormalities or signs of hydrops was detected. Family declined genetic testing and opted to continuation of the pregnancy. Antenatal MRI showed hypertrophy of soft tissues with multiple vascular channels in interfacial and intermuscular planes on T1 and T2 weighted imaging (Fig 4). After birth, the neonate exhibited asymmetric hypertrophy of the right lower limb, accompanied by prominent dark reddish superficial capillary malformations on the skin (Fig 5). Postnatal MRI revealed an infiltrating vascular lesion involving the subcutaneous, intramuscular and intermuscular planes. These findings confirmed significant congestion in the right lower limb and led to a definitive diagnosis of Klippel Trenaunay Syndrome

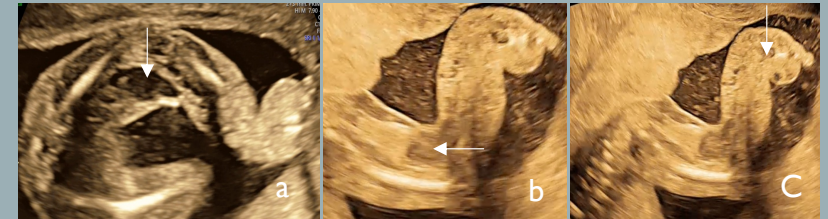


Fig 1. Diffuse heterogeneous lesion with tortuous channels and cystic spaces (arrows) seen diffusely involving the subcutaneous tissues and muscles of the right lower limb (thigh, leg and foot (a,b,c).

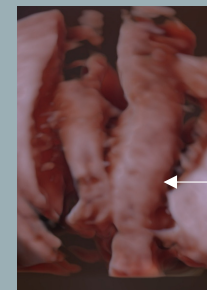


Fig 2. The effected right lower limb (arrow) is markedly thicker and oedematous as compared to the normal left lower limb

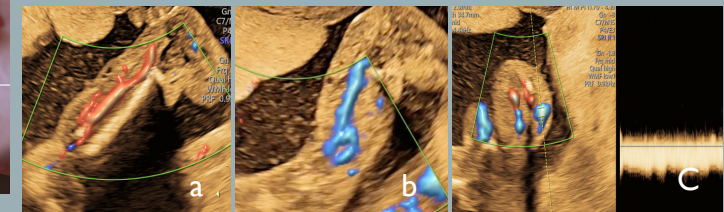


Fig 3. Power Doppler imaging demonstrated spontaneous flow within the dilated anechoic channels (a, b) Spectral doppler showing continuous flow indicative of venous lacunae (c)

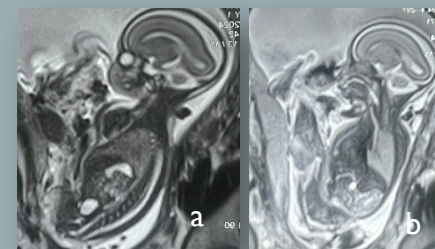


Fig 4. Antenatal MRI showing multiple vascular channels in interfacial and intermuscular planes on T1 and T2 weighted imaging



Fig 5. Postnatal image
Hypertrophied lower limb with prominent dark reddish superficial capillary malformations on the skin.